



Williams, A. G., McNally, J., Gaskin, J., & Chakkarapani, E. (2018). Preterm Infant with Congenital Tracheal Diverticulum in the Presence of Esophageal Atresia and Tracheoesophageal Fistula. *Journal of Neonatal Surgery*, 7(4), [44]. <https://doi.org/10.21699/jns.v7i4.741>

Peer reviewed version

License (if available):
CC BY

Link to published version (if available):
[10.21699/jns.v7i4.741](https://doi.org/10.21699/jns.v7i4.741)

[Link to publication record in Explore Bristol Research](#)
PDF-document

This is the author accepted manuscript (AAM). The final published version (version of record) is available online via EImed Publishers at <http://www.jneonatalurg.com/index.php/jns/article/view/741> at [insert hyperlink] . Please refer to any applicable terms of use of the publisher.

University of Bristol - Explore Bristol Research

General rights

This document is made available in accordance with publisher policies. Please cite only the published version using the reference above. Full terms of use are available:
<http://www.bristol.ac.uk/red/research-policy/pure/user-guides/ebr-terms/>

Preterm infant with congenital tracheal diverticulum in the presence of oesophageal atresia and tracheo-oesophageal fistula

Glover Williams A¹, McNally J², Gaskin J³, Chakkarapani E⁴

1. Paediatric Service, Royal United Hospitals Bath NHS Foundation Trust
2. Paediatric Surgery, Bristol Royal Hospital for Children
3. ENT, Bristol Royal Hospital for Children
4. Neonatal Intensive Care Unit, St Michael's Hospital, Bristol

Corresponding author:

Alessandra Glover Williams

Paediatric Service, Royal United Hospitals Bath NHS Foundation Trust
Dept of Paediatrics,
Royal United Hospital Bath,
Combe Park,
Bath
BA1 3NG
Email: alessandra.glover1@nhs.net
Phone: 07702093898

Abstract

A preterm infant born at 32+4 weeks gestation had a tight, primary anastomosis of the oesophageal atresia and closure of a tracheo-oesophageal fistula on day one of life. He remained ventilated for six days post operatively and following extubation needed continuous positive airway pressure due to respiratory insufficiency and increased work of breathing. Postoperative course was complicated by right hemi-diaphragmatic paresis and acute life threatening episodes of deep desaturation and bradycardia requiring high-pressure, non-invasive positive pressure ventilation. Definitive diagnosis of congenital tracheal diverticulum and severe tracheomalacia was made with computed tomography assisted by three dimensional reconstruction and confirmed with microlaryngoscopy and bronchoscopy. Investigation of this rare diagnosis and its subsequent management is discussed in the report with reference to the literature.

Introduction

Following oesophageal atresia (OA) repair and closure of tracheoesophageal fistula (TOF) common early complications include anastomotic leak, gastro-oesophageal reflux, anastomotic stricture and missed proximal TOF.[1,2] One of the rare, late complications, tracheal diverticulum, occurs at the site of ligation of fistula and presents at later life around a median age of 12 months with repetitive respiratory symptoms and usually diagnosed using bronchoscopy.[3] There are no previous reports of congenital tracheal diverticulum associated with TOF/OA in the literature. Here we describe a rare anomaly of congenital tracheal diverticulum associated with OA/TOF presenting early at two weeks of age in a preterm infant. We describe a novel approach to investigation using computed tomography (CT) and 3D reconstruction along with microlaryngoscopy and bronchoscopy in the neonatal period and subsequent management. Congenital tracheal diverticulae are a poorly recognised finding in OA/TOF and can be asymptomatic particularly if distal in location, but in the presence of severe tracheomalacia, they may present early with respiratory symptomology.[4]

Case Report

The patient was born by spontaneous vaginal delivery at 32+4 weeks gestation after a late antenatal diagnosis of OA. He had a tight, primary anastomosis of the OA on day one of life and closure of a TOF. There was no microlaryngoscopy and bronchoscopy (MLB) undertaken prior to repair. He remained ventilated for six days post operatively and following extubation needed continuous positive airway pressure (CPAP) due to respiratory insufficiency and increased work of breathing. Postoperative chest radiograph showed right hemi-diaphragmatic paresis.

He soon developed acute life threatening episodes (ALTEs) of deep desaturation and bradycardia requiring high-pressure, non-invasive positive pressure ventilation. These episodes persisted for weeks and occurred when he was unsettled or straining. Medical management of potential gastro-oesophageal reflux had no effect and these episodes were not associated with seizure activity on the electroencephalograph at six weeks of age. MLB was undertaken and diagnosed severe, lower tracheomalacia. Prior to and during the procedure he suffered from a cardiorespiratory collapse requiring resuscitation, which prematurely terminated the investigation and he returned from theatre intubated.(Fig 1)

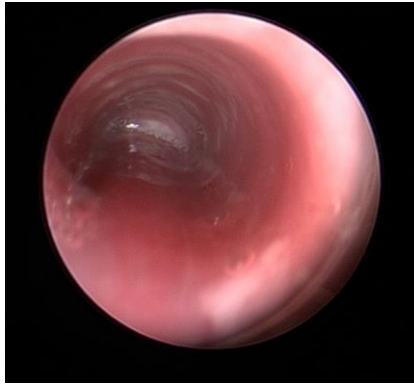


Figure 1. MLB view of tracheomalacia

Following this he had a CT chest with contrast at 4 weeks of age to exclude a vascular ring or external compressing structure. The CT found a posterior tracheal diverticulum with the origin just proximal to the carina, lower than the TOF/OA repair. The distal end of the diverticulum ended below the carina, dividing into two blind ends like those anatomically expected from bronchiolar division.(Fig 2) The aetiology of the pouch was determined to be congenital tracheal diverticulum.

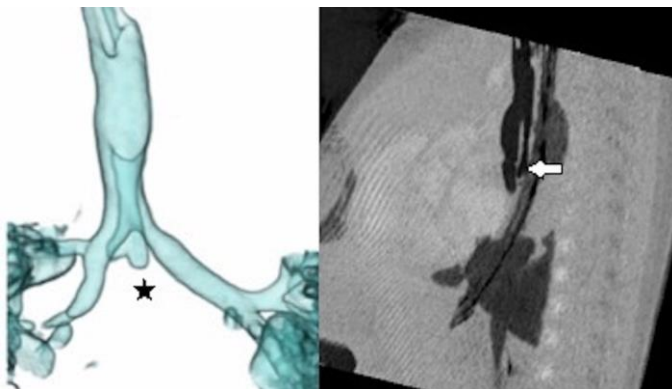


Figure 2. 3D CT reconstruction of the tracheal diverticulum (starred) and 2D CT in the saggital plane (arrowed)

He continued to suffer from ALTEs whilst intubated. Ten days following the CT, he had a second MLB, which confirmed presence of the diverticulum with the entrance 1cm proximal to the carina.(Fig 3) As part of the MLB he was intubated past the diverticulum; this stabilised his airway and the ALTEs resolved. Upper gastrointestinal contrast confirmed only mild stenosis of the oesophageal repair, without a fistula.

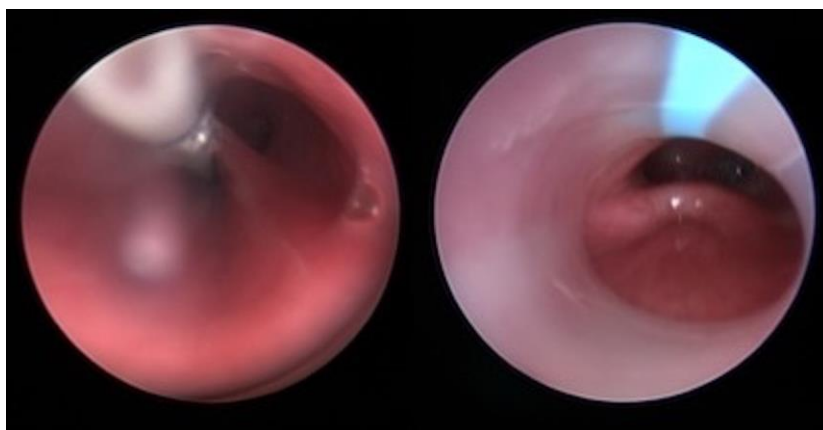


Figure 3. MLB view of the diverticulum

Definitive treatment involved surgical resection of the diverticulum with aortopexy. Further imaging ruled out VACTERL sequence. Brain magnetic resonance imaging (MRI) undertaken to investigate the extent of hypoxic damage from multiple episodes of resuscitation showed haemosiderin deposition over the left frontal convexity and haemorrhage in the cerebellum; there was no evidence of hypoxic brain injury in the diffusion weighted imaging, T1 and T2 sequences. He did not require tracheostomy and is breathing without any respiratory support in room air.

Discussion

OA and TOF affect 1:300-4500 live births, most commonly in combination.[1] The majority are repaired in the first days of life by primary anastomosis if the oesophageal gap will allow. It is preferable to extubate onto air and avoid re-intubation or positive pressure ventilation to protect the anastomosis and avoid anastomotic leak. When a patient is born prematurely the chance of extubation onto room air without non-invasive support is lower due to the lung immaturity, as with our patient.

Tracheal diverticulum can also present with airway compromise and difficult ventilation.[2] Tracheal diverticulum is best diagnosed with dynamic three phase bronchoscopy, however the airway instability of our patient hindered attempts. It is potential that pre-operative MLB could have diagnosed the presence of both a congenital diverticulum along with that of a TOF, however the tracheomalacia would still have proved problematic postoperatively and required further surgical intervention.

Although multi-dimensional CT with 3D reconstruction has been used in adults to diagnose tracheal diverticulum, this is the first report of use in a preterm neonate to diagnose tracheal diverticulum. As mentioned above, the bifurcation of the diverticulum as seen in the 3D CT reconstruction suggests the potential development of bronchioles from the structure, and thus that the diverticulum was congenital and not acquired. The length of the diverticulum

also supports the congenital diagnosis, along with the fact that the OA closure was high and a TOF would not extend so low or indeed to beyond the carina, as the diverticulum does. Histology of the diverticulum was consistent with a tracheal pouch, however the tissue had considerable diathermy artefact and so could not be considered diagnostic. We accept that this is a limitation along with the unfortunate absence of MLB confirmation, but feel that the bifurcating structure on imaging is significantly suggestive of a tracheal origin.

The understanding of the embryogenesis of this combination is poorly understood however, the embryogenesis of TOF/OA alone also remains poorly understood. Currently the most popular theories of TOF/OA formation consist of a failure of separation of the dorsal and ventral anterior foregut into the gastrointestinal and respiratory components respectively at around 3-5 weeks gestation.[5] The drive for this failure appears to be dependent upon the molecular expression of growth factors, however the growth factors involved remains to be established. Congenital tracheal diverticulum is believed to occur from a defect of endodermal differentiation and tracheal cartilage formation during the sixth week of development.[6] It is plausible that altered growth factors resulting in TOF/OA formation, could also impact the posterior wall formation of the trachea, resulting in diverticulum, but this is purely speculative.

Despite progressive improvement in the surgical management of OA/TOF, the majority of patients experience long-term structural and functional oesophageal and tracheal complications such as tracheomalacia.[7] Our patient had severe tracheomalacia, which was treated with aortopexy. Patients with severe tracheomalacia are more likely to experience ALTEs and require aortopexy or tracheostomy formation.[8] Tracheomalacia is evaluated most commonly with MLB or fluoroscopy. In a case series of patients with OA, 18.9% developed severe tracheomalacia at a median of 18 days post surgery.[9] While 20% of these patients died, others went on to have aortopexy, fundoplication or medical management. Patients managed with aortopexy had shorter length of hospital stay and fewer readmissions for respiratory issues at two-year follow-up.[9] This suggests that timely aortopexy for severe tracheomalacia might be the best treatment option. Posterior tracheopexy is also emerging through the Oesophageal and Airway Treatment team at Boston Children's Hospital as a primary or secondary treatment option for tracheomalacia associated with OA. Symptom resolution has been found to be significant, as has ventilator dependence, compared with primary and secondary tracheopexy boasting comparable results.[10] Only 9.2% of 98 patients required reoperation such as aortopexy.

Conclusion

There are many early and late complications associated with OA/TOF repair and a multi-disciplinary approach to diagnostic investigation is key, potentially

with MLB prior to repair to prevent unexpected developments postoperatively as much as possible.

Congenital tracheal diverticulum in the presence of OA/TOF is a rare diagnosis, not previously comprehensively addressed in the literature and provides significant challenges in management in the presence of tracheomalacia.

Bronchoscopy and CT is the gold standard of investigation for tracheal diverticulum and indication for surgical management is assessed on severity of symptoms.

Currently management of severe tracheomalacia consists of aortopexy or posterior tracheopexy, with surgical developments questioning the case for preventative primary tracheopexy at the time of TOF/OA repair.

References

1. Alberti D, Boroni G, Corasaniti L et al. [Esophageal atresia: pre and post-operative management](#). *J Matern Fetal Neonatal Med*. 2011 Oct;24 Suppl 1:4-6.
2. Gibreel W, Zendejas B, Antiel, RM et al. Swallowing Dysfunction and Quality of Life in Adults With Surgically Corrected Esophageal Atresia/Tracheoesophageal Fistula as Infants: Forty Years of Follow-up. *Ann Surg*. 2017;266:305–310.
3. Shah AR, Lazar EL, Atlas AB. Tracheal diverticula after tracheoesophageal fistula repair: case series and review of the literature. *J Pediatr Surg*. 2009 Nov;44(11):2107-11. doi: 10.1016/j.jpedsurg.2009.04.036.
4. Johnson LB, Cotton RT, Rutter MJ. Management of symptomatic tracheal pouches. *Int J Pediatr Otorhinolaryngology*. 2007; 71: 527—531.
5. Iannides A, Copp A. Embryology of oesophageal atresia. *Semin Pediatr Surg*. 2009 Feb; 18(1):2-11.
6. Tanaka H, Mori Y, Kurokawa K et al. Paratracheal air cysts communicating with the trachea: CT findings. *J Thorac Imaging*. 1997 Jan; 12(1):38-40
7. Kovesi T. Long-term respiratory complications of congenital esophageal atresia with or without tracheoesophageal fistula: an update. *Dis Esophagus*. 2013 May-Jun;26(4):413-6. doi: 10.1111/dote.12061.
8. Shah R, Varjavandi V, Krishnan U. Predictive factors for complications in children with esophageal atresia and tracheoesophageal fistula. *Dis Esophagus*. 2015 Apr;28(3):216-23. doi: 10.1111/dote.12177.
9. Kay-Rivest E, Baird R, Laberge JM et al. Evaluation of aortopexy in the management of severe tracheomalacia after esophageal atresia repair. *Dis Esophagus*. 2015 Apr;28(3):234-9. doi: 10.1111/dote.12179. Epub 2014 Jan 22.
10. Sheih HF, Smithers CJ, Hamilton TE et al. Posterior tracheopexy for severe tracheomalacia associated with esophageal atresia (EA): Primary Treatment at the time of initial EA repair versus secondary treatment. *Front Surg*. 2017; 4: 80.